Brief Communication

Double Papillary Fibroelastoma of the Aortic Valve

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A case of asymptomatic aortic valve fibroelastoma was diagnosed by transthoracic and transesophageal echocardiography in a 77-year-old man. A 2nd tumor was found at surgery. Both fibroelastomas were removed surgically without aortic valve replacement. During surgical excision of a cardiac fibroelastoma, the entire area surrounding the lesion should be inspected carefully for additional lesions. Such lesions can be missed on echocardiographic evaluation. ***(Tex Heart Inst J 2004;31:448-9)***

Cardiac papillary fibroelastomas are the third-most-common benign tumors of the heart, after myxomas and lipomas, and they are the most common valvular neoplasm.[1] Clinical manifestations include stroke, myocardial infarction, pulmonary embolism, and, less commonly, transient cerebral ischemic attack, angina, congestive heart failure, syncope, and sudden death.[2] These tumors can occur as isolated lesions or in association with other cardiac diseases. They are usually solitary tumors, although they appear at multiple sites in 7.5% of cases.[3] This incidence could be underestimated, because sometimes only a thorough surgical inspection reveals the smaller, additional lesions.[4,5]

Case Report

In March 2003, a 77-year-old man was referred to our department for evaluation and treatment of an aortic valve tumor. Relevant medical history included systemic hypertension and atrial fibrillation with a low ventricular rate that required permanent pacing. For this reason, the patient was taking warfarin at the time of admission. Diffuse, severe atherosclerosis was evidenced by carotid and femoral artery stenoses and ascending aorta calcifications. Transesophageal echocardiography (TEE) showed a small mobile mass that was attached to the left coronary cusp by a short thin stalk (Fig. 1). Coronary angiography revealed occlusion of a nondominant right coronary artery. At surgery, the ascending aorta was examined by TEE, and a disease-free area was selected for cannulation and cross-clamping. The left coronary cusp tumor was found to be pedunculated, with a diameter of 5×10 mm. On the noncoronary cusp, we found a 2nd small mass (2×4 mm) with a broad implantation base (Fig. 2). Both lesions clearly originated from the leaflet-free edge and could be excised simply. Intraoperative TEE documented trivial aortic valve regurgitation that had been present preoperatively.

The patient's postoperative course was uneventful. Histopathologic examination confirmed the diagnosis of cardiac papillary fibroelastoma in both lesions. When the patient was last seen in January 2004, he was in New York Heart Association functional class I, with mild aortic valve regurgitation and no evidence of tumoral regrowth.

Discussion

The incidence of papillary fibroelastoma is less than 10% of all cardiac tumors.[2] In some cases, cardiac fibroelastomas are considered to be congenital; however, in most patients they are acquired.[1] These tumors occur equally often in males and females. They were first described as incidental findings at autopsy, with a prevalence of 0.002% to 0.33%, or at surgery.[2] The clinical significance of papillary fibroelastomas has been debated extensively; such controversy persists even when

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the risk of cerebral or coronary embolization indicates surgical excision.

Papillary fibroelastomas originate from the valves or the endothelial surface of the heart. In a recent review of 410 reported cardiac papillary fibroelastomas, 84% arose from cardiac valves.[6] Another study reported cardiac papillary fibroelastomas to arise from aortic, mitral, tricuspid, and pulmonic valves in 35%, 25%, 17%, and 13% of cases, respectively.[1] These tumors are small, avascular masses, which, in 91% of cases, are single lesions. Frequently, they are mobile with a thin stalk (43%) and have a mean diameter of 3×10 mm.[7] They have multiple papillary fronds that cause them to resemble sea anemones. Histologically, papillary fibroelastomas consist of a central core of dense connective tissue, surrounded by a layer of loose connective tissue and covered by hyperplastic endothelial cells.

Detection of papillary fibroelastomas has increased with the use of echocardiography. Echodensity of the tumor's central collagen core strongly supports the diagnosis and allows differentiation from other intracardiac tumors, vegetations, or mural thrombi.[7] Echocardiographic examination does not reliably reveal multiple lesions, although it is the best diagnostic tool for papillary fibroelastomas. Coronary angiography is not advisable because of the fragile nature of the tumor.

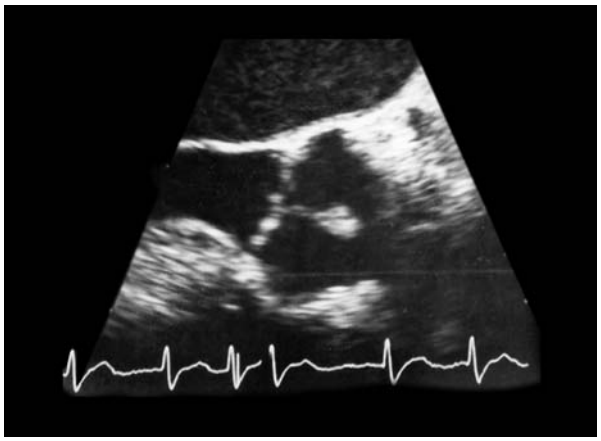

Fig. 1 *Preoperative transesophageal echocardiogram shows only the larger cardiac papillary fibroelastoma adhering to the free edge of the left coronary cusp.*

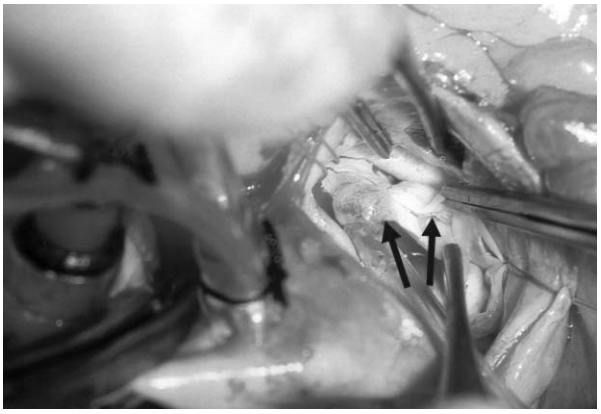

Fig. 2 *Operative view shows a secondary mass (arrows) arising from the noncoronary cusp.*

The 1st surgical resection of a cardiac papillary fibroelastoma was performed by Lichtenstein in 1979.[8] The presence of a such a tumor is an indication for surgical resection, because even small lesions (3–4 mm) can cause life-threatening complications. After tumor excision, aortic leaflets can be restored with a homograft aortic cusp or an autologous pericardial patch. Tumor resection alone may be sufficient, especially when the mass arises from the cusp-free edge. Normal function and structure of the aortic valve can usually be restored.

It is rare to find multiple lesions of the aortic valve, in part because additional lesions may be missed on TEE, as in our patient. In many cases, a 2nd lesion is detected only upon surgical inspection.[4,5] Therefore, the entire heart should be assessed as thoroughly as possible during surgery. Smaller lesions may be attached beneath the leaflet surface. Moreover, multiple cardiac papillary fibroelastomas can involve separate valves or both the left and right cavities.[3] Additional lesions most likely represent different stages of the same tumor.[9] Recurrence rates of papillary fibroelastomas after surgical excision have not been reported, but careful follow-up is necessary.

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